May 2017 Imaging Case of the Month

Prasad M. Panse, MD and Michael B. Gotway, MD

Department of Radiology
Mayo Clinic Arizona
Scottsdale, Arizona USA

Clinical History: A 32-year-old man presented for routine physical examination. His past medical history is unremarkable and the physical examination and basic laboratory data were within normal limits.

A frontal chest radiograph (Figure 1) was performed.

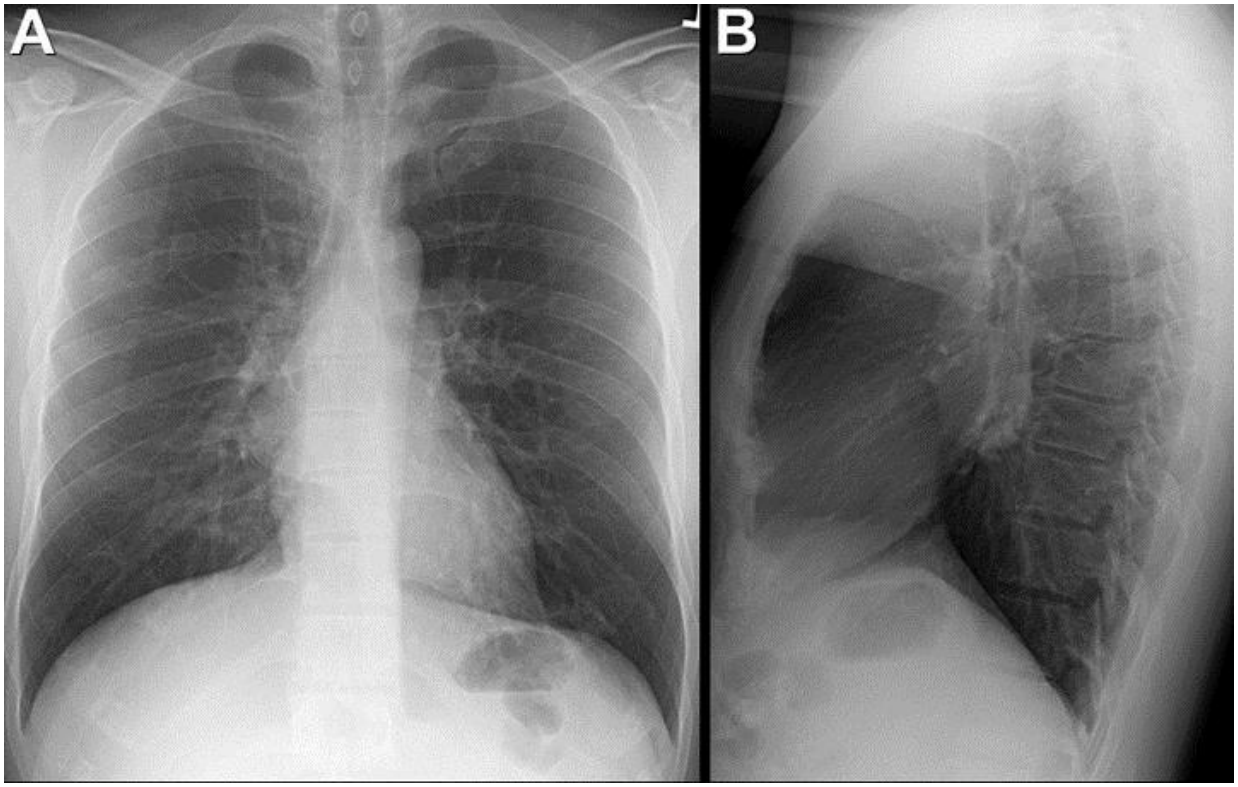

Figure 1: Frontal (A) and lateral (B) chest radiography.

Which of the following statements regarding the chest radiograph is ***most accurate***?

1. The frontal chest radiograph shows an abnormal mediastinal contour
2. The frontal chest radiograph shows basal predominant fibrotic abnormalities
3. The frontal chest radiograph shows large lung volumes with a cystic appearance
4. The frontal chest radiograph shows multifocal small pulmonary nodules
5. The frontal chest radiograph shows no abnormal findings

Correct!

1. The frontal chest radiograph shows an abnormal mediastinal contour

The frontal chest radiograph shows normal lung volumes and clear lungs; no pleural abnormality is present. There is no evidence of cystic lung disease or basal fibrotic abnormalities. An abnormal mediastinal contour is present on both the frontal and lateral chest radiographic projections (Figure 2).

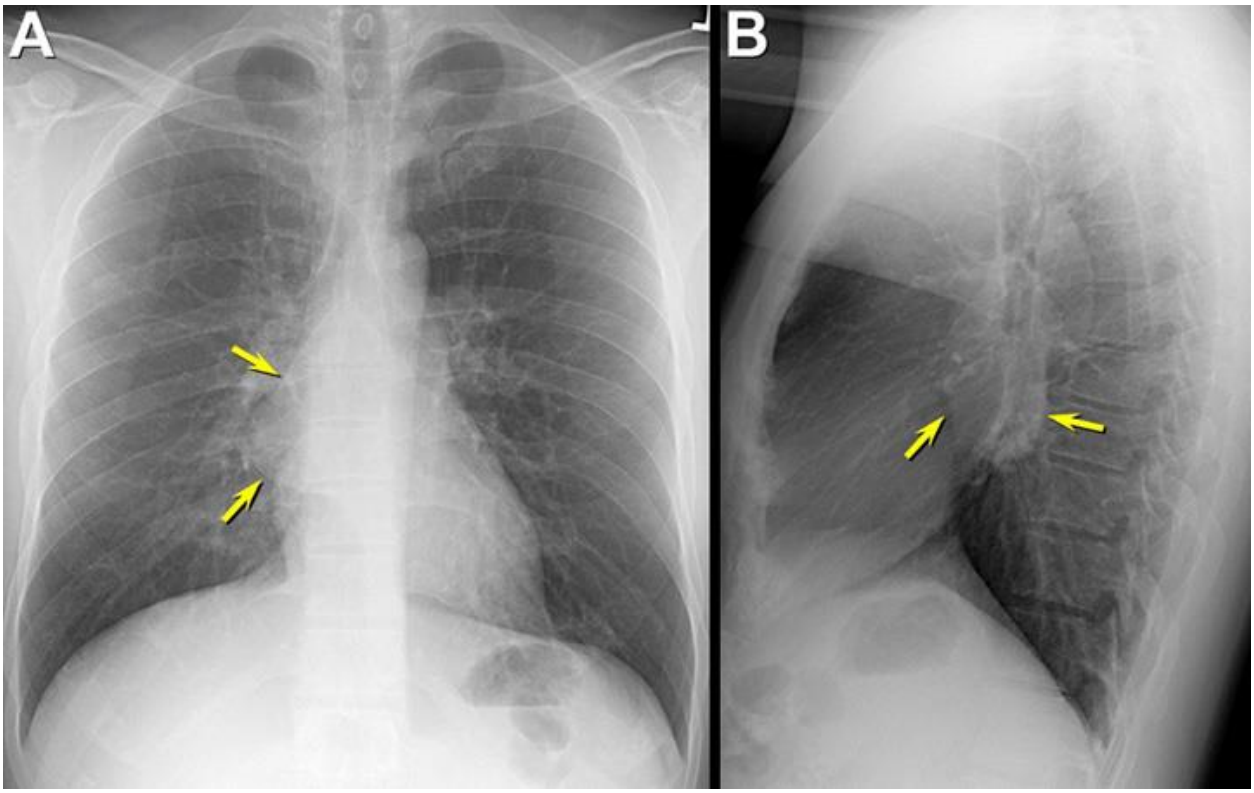

Figure 2. Frontal (A) and lateral (B) chest radiography shows an abnormal mediastinal contour creating a convex “bulge” in the ayzgoesophageal recess (arrows, A). The abnormal mediastinal contour is clearly visible in the infrahilar window region on the lateral chest radiograph (arrows, B).

Regarding the frontal chest radiograph (Figure 1) abnormality, which of the following statements regarding the chest radiograph is ***most accurate***?

1. The abnormal mediastinal contour affects the aorto-pulmonary window
2. The abnormal mediastinal contour affects the ayzgoesophageal recess
3. The abnormal mediastinal contour affects the pre-aortic space
4. The abnormal mediastinal contour affects the right para-aortic stripe
5. The abnormal mediastinal contour affects the right paratracheal stripe

Correct!

2. The abnormal mediastinal contour affects the ayzgoesophageal recess

The frontal chest radiograph (Figure 1A) shows an abnormal mediastinal contour bulge that causes an outward convexity of along the mid-portion of the ayzgoesophageal recess. Normally the ayzgoesophageal recess is a mediastinal interface created by contact of the medial right lung with the right mediastinum extending cranially from the arch of the azygos vein caudally to the diaphragm. The mediastinum in this location typically contains the azygos vein, esophagus, small lymph nodes, and mediastinal fat, the former two contributing to the name of the ayzgoesophageal recess, with the azygos vein (and pleural covering the thoracic spine) often creating the posterior border, and the esophagus the medial border, of the ayzgoesophageal recess, although the anatomy may vary substantially from patient to patient. Typically the ayzgoesophageal recess describes a concave configuration with the adjacent right lung, or often a gentle "S"-shaped curve may be seen (Figure 3).

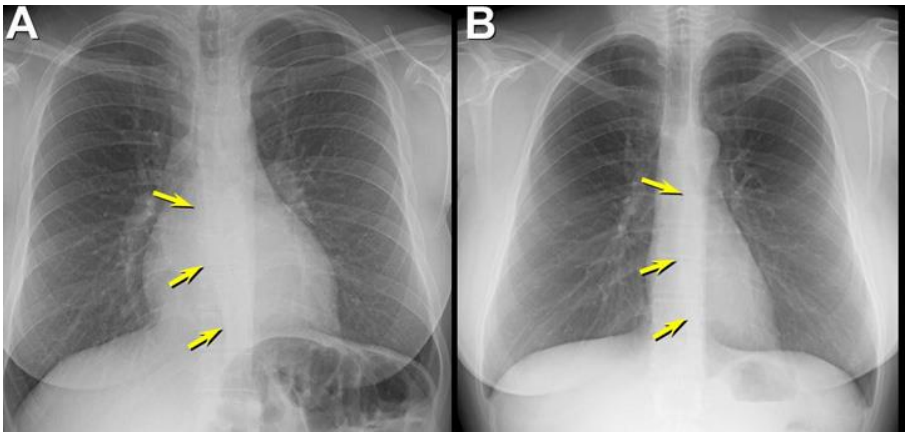

Figure 3. Frontal chest radiographs in 2 separate patients showing the normal interface created between the medial right lung and the medial right mediastinal region, referred to as the ayzgoesophageal recess. In these patients, the ayzgoesophageal recess describes a gentle, "S"-shaped curve.

Abnormalities of the ayzgoesophageal recess present as outward, convex "bulges" of this line, which may be noticeably focal in some circumstances. The right paratracheal stripe and right para-aortic stripe both appear normal in this patient. The aorto-pulmonary window and pre-aortic spaces also both appear normal and both are left-sided structures.

Which of the following is the ***most appropriate*** next management step for this patient?

1. ^{18}FDG – PET scan
2. Contrast enhanced thoracic CT
3. Echocardiography
4. Lateral decubitus chest radiography
5. Ventilation – perfusion scintigraphy

Correct!
2. Contrast enhanced thoracic CT

Contrast-enhanced thoracic CT is the most appropriate selection among the choices listed above. The injection of intravenous contrast, as opposed to an unenhanced thoracic CT, is generally preferred when the indication for thoracic CT is for evaluation of a mediastinal mass. Intravenous contrast is essential if the mass due to vascular abnormality, and when masses do not arise from vascular structures, intravenous contrast is nevertheless still very useful because intravenous contrast administration allows for optimal assessment of the relation of the mass to surrounding vascular structures, which is important both for pre-operative assessment and determining the biologic behavior of the lesion. ^{18}FDG – PET scan could prove useful for evaluation of the mass, but generally ^{18}FDG – PET scan is most rewarding when obtained for further characterization of indeterminate findings at thoracic CT, rather than immediately following chest radiography. Lateral decubitus chest radiography is useful for assessment of possible pleural effusion (for evaluation of the dependent thorax), or for assessment of pneumothorax (when attention is focused on the non-dependent thorax); neither situations are relevant here. Ventilation – perfusion scintigraphy is most commonly employed for the assessment of acute or chronic thromboembolic disease, or occasionally for determination of differential pulmonary perfusion prior to thoracic surgery or for assessment of systemic shunting; none of these conditions are relevant to this outpatient with an incidentally detected, incompletely evaluated mediastinal mass. Echocardiography could potentially visualize the lesion and provide some characterization of the mediastinal mass, but it is likely that this lesion resides outside the heart and would not be the focus of an echocardiogram and probably would be incompletely visualized.

The patient subsequently underwent unenhanced thoracic CT (Figure 4).

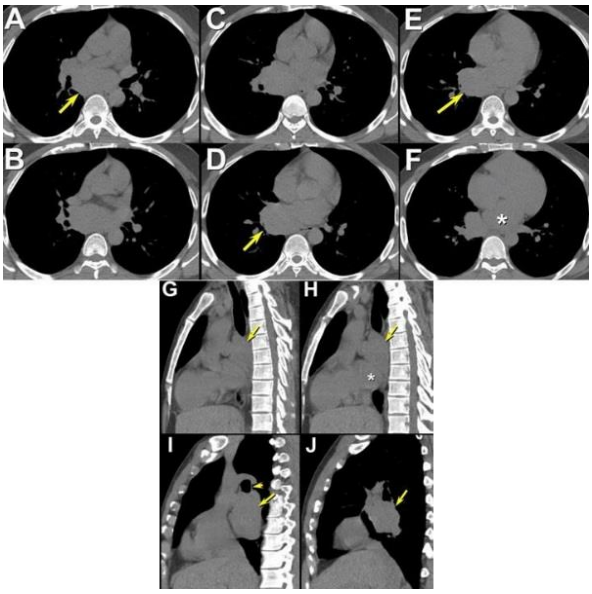

Figure 4. Representative images from unenhanced axial (A-F) and sagittal (G-H) CT.

Regarding this examination, which of the following is ***correct***?

1. The thoracic CT shows a destructive chest wall mass
2. The thoracic CT shows a mass arising from the pleura
3. The thoracic CT shows a mildly hyperattenuating mass in the subcarinal region extending inferiorly into the azygoesophageal recess
4. The thoracic CT shows enlargement of the right pulmonary artery
5. The thoracic CT shows multifocal mediastinal and peribronchial lymph node enlargement

Correct!

6. The thoracic CT shows a mildly hyperattenuating mass in the subcarinal region extending inferiorly into the azygoesophageal recess

The thoracic CT shows normal pleural surfaces and the chest wall appears normal; no evidence of bone destruction or chest wall mass is present. A mediastinal mass, representing the lesion seen at chest radiography, is present, located in the subcarinal region. There is no evidence of either peribronchial or mediastinal lymph node enlargement outside the subcarinal region. The right pulmonary artery is normal in size, but is displaced laterally by the azygoesophageal mass.

The thoracic CT was inadvertently protocoled as an unenhanced examination, and therefore characterization of the lesion was incomplete. Attenuation measurements of the lesion shows that the lesion ranged in density from 38- 46 HU, which is consistent with complex fluid within a cyst, but could potentially be seen with a solid mass (Figure 5).

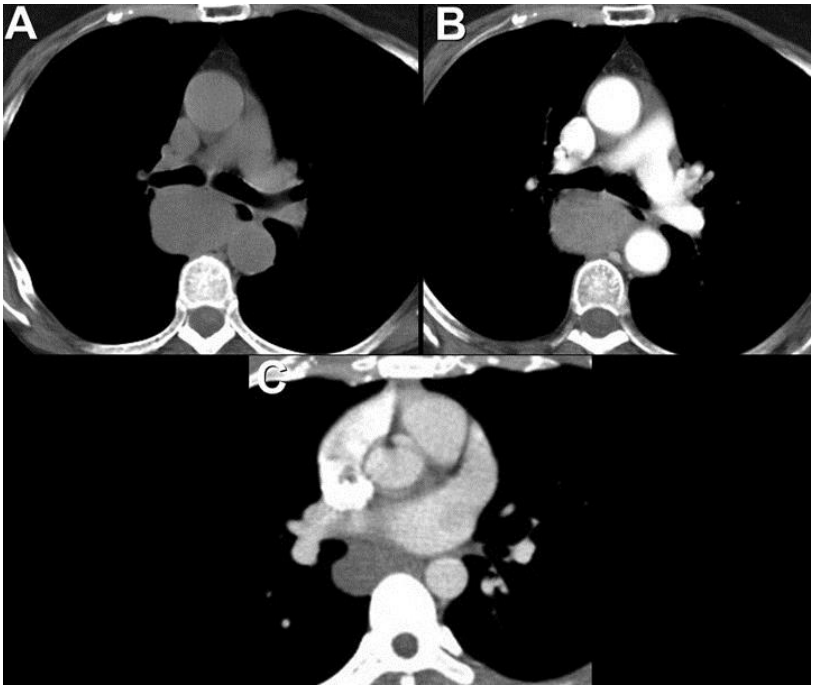

Figure 5. Axial unenhanced (A) and enhanced (B) thoracic CT in a patient with esophageal leiomyoma shows a lesion just posterior to the carina that is mildly hyperattenuating on the unenhanced image (A); attenuation coefficients measured about 45 HU. Following contrast administration (B), the lesion shows enhancement, confirming its solid nature. Axial enhanced image in a different patient (C) with surgically-proven esophageal leiomyoma shows a homogeneous mass that measures only 52 HU following intravenous contrast administration; a previous unenhanced study (not shown) revealed that this lesion measured about 32 HU prior to intravenous contrast administration. These cases underscore the need for unenhanced and enhanced imaging for proper characterization of some mediastinal masses.

Which of the following is the ***most appropriate next step*** for the management of this patient?

1. ^{18}FDG – PET scan
2. Endoscopic ultrasound with biopsy
3. Follow up enhanced thoracic CT in 3 months
4. Repeat unenhanced and enhanced thoracic CT
5. Unenhanced and enhanced thoracic MR

Correct!
5. Unenhanced and enhanced thoracic MR

Unenhanced and enhanced imaging is integral to characterization of a number of mediastinal lesions, particularly mediastinal cysts, which are among the more common causes of mediastinal masses, particularly when incidentally detected in younger patients. In this patient, unenhanced followed by enhanced MR provides the unique ability to characterize this lesion without the use of ionizing radiation, and therefore obviates the need for repeat unenhanced and enhanced thoracic CT. ^{18}FDG – PET scan could prove useful for evaluation of the mass, but the lesion should be further characterized with unenhanced and enhanced imaging without further exposure of this young patient to ionizing radiation. Follow up thoracic CT is occasionally useful for characterization of small, indeterminate pulmonary opacities, particularly when malignancy is considered, but would not play a reasonable role for the assessment of a nearly 6 cm mediastinal mass. Endoscopic ultrasound could prove useful for characterizing this lesion given its proximity to the esophagus (or using endobronchial ultrasound, given the proximity of the lesion to the tracheal carina), but biopsy should be avoided- if the lesion is indeed a cyst, endoscopy with biopsy could result in spillage of the cyst contents into the mediastinum and also runs the risk of introducing infection into a previously closed space.

To further characterize the lesion discovered at CT scanning, the patient was underwent thoracic MR (Figure 6).

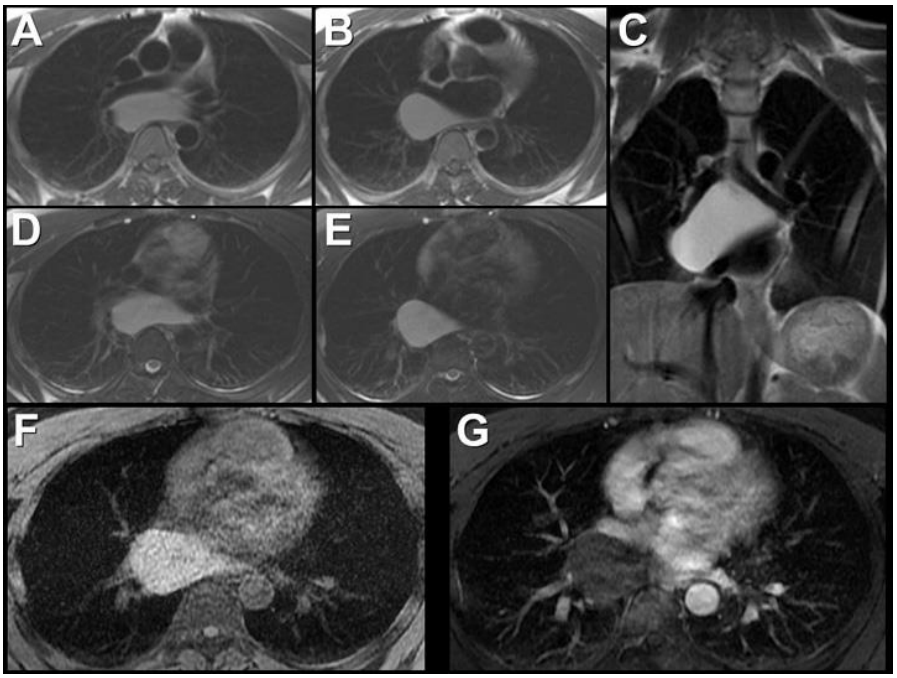

Figure 6. Representative images from thoracic MRI. T1-weighted axial (A and B) and coronal (C) images. Axial T2-weighted imaging with fat saturation (a fluid-sensitive sequence; D and E). Axial unenhanced (F) and enhanced (G) gradient echo imaging.

Which of the following regarding this MR examination is ***correct***?

1. The thoracic MR reveals additional sites of abnormality, suggesting a systemic process
2. The thoracic MR shows that the lesion contains fat
3. The thoracic MR shows that the lesion does ***not*** enhance significantly
4. The thoracic MR shows that the lesion is solid
5. The thoracic MR shows the same mass at CT, but adds little additional information to that already available with CT

Correct!

3. The thoracic MR shows that the lesion does *not* enhance significantly

The thoracic MR shows the same azygoesophageal recess mass seen at CT, but adds additional information regarding the azygoesophageal recess lesion's tissue components. The azygoesophageal recess lesion does show increased signal intensity on the T1-weighted images, which is common with fat [note the hyperintense signal of subcutaneous fat on these images]; however, the lesion continues to show hyperintense signal on T2-weighted MR sequencing employing fat saturation, indicating that the internal hyperintensity of the lesion is *not* due to fat (see Figures 6D and E). *Fat saturation* refers to MR techniques designed to eliminate signal from fat. When a lesion shows high signal on T1-weighted imaging, suggesting the possibility of intralesional fat, if that signal disappears when using fat-saturation techniques, then one can be relatively certain that the hyperintensity of the lesion on T1-weighted imaging is not due to the presence of intralesional fat; rather, the increased signal could be due to other causes of hyperattenuation on T1-weighted imaging, particularly hemorrhage or proteinaceous fluid. The increased signal on the fluid-sensitive images suggests the possibility of a cyst, but some solid lesions can display this behavior as well, so the lesion cannot be characterized as a cyst on this sequence alone. However, the images obtained before and after intravenous contrast administration, which show no evidence of enhancement, combined with the fluid signal on the fluid-sensitive T2-weighted sequences, are consistent with a cystic mediastinal lesion in the azygoesophageal recess l. The thoracic MR study shows no other sites of abnormality.

Which of the following represents the ***next most appropriate step*** for the evaluation of this patient?

1. ^{18}FDG-PET
2. ^{68}Ga-citrate scintigraphy
3. Mediastinoscopy
4. Offer surgical excision
5. Percutaneous transthoracic fine needle aspiration biopsy

Correct!
4. Offer surgical excision

The imaging features are consistent with a particular diagnosis, and invasive testing is not necessary. Therefore, percutaneous transthoracic fine needle aspiration biopsy and mediastinoscopy are not the correct answers. The risk of spillage of the cystic content during percutaneous transthoracic fine needle aspiration biopsy is unacceptably high whereas the likelihood of obtaining a histopathological diagnosis is low for this cystic, isolated, indolent-appearing, and incidentally detected lesion. The lesion may also be too caudally located in the subcarinal space to be reached via mediastinoscopy. While ^{18}FDG-PET could potentially be a useful procedure for the evaluation of mediastinal lesions, as it has the ability to assess for metabolic activity within the lesion as well as the ability to detect potential sites of disease elsewhere within and outside the thorax, the lesion has already been well-characterized by cross sectional imaging. The lack of tracer utilization within the lesion would simply reinforce the impression of a benign abnormality, whereas increased tracer utilization would most likely prompt incorrect consideration of an aggressive process for this lesion. ^{68}Ga-citrate scintigraphy would not provide addition useful information in this patient. Among the choices listed, offering surgical resection is reasonable. The lesion could be observed, given that the patient apparently is not exhibiting symptoms related to the presence of the lesion, but the sheer size of the lesion mandates some consideration for surgical resection.

Based on the data thus far, which of the following represents the ***most likely diagnosis*** for this patient?

1. Bronchogenic / foregut duplication cyst
2. Isolated azygoesophageal recess metastasis
3. Müllerian duct (Hattori) cyst
4. Pericardial cyst
5. Thoracic duct cyst

Correct!
1. Bronchogenic / foregut duplication cyst

As noted previously, the cross sectional imaging findings indicate an indolent-appearing, incidentally discovered cyst in the ayzgoesophageal recess. Occasionally some malignancies, such as renal cell malignancies and squamous cell carcinomas, can occasionally appear necrotic, even cystic, and can manifest as isolated mediastinal lesions. However, this patient has no active or previous malignancy, and even prominently cystic or necrotic carcinoma metastases usually show some solid, enhancing areas; the latter are lacking in this patient. The lesion's location is unusual for pericardial cysts (typically anteriorly located in the right > left cardiophrenic angle), Müllerian duct (aka Hattori) cysts (typically posterior- superior mediastinum in women), and thoracic duct cysts (posterior mediastinum, connected to the thoracic duct). However, the lesion's location in the azygoesophageal recess, near the carina, is a common location for a bronchogenic / foregut duplication cyst, which also is the most common cause of a cystic middle mediastinal lesion.

Further questioning disclosed that the patient had undergone a right thoracotomy (see Figure 1; note the thoracotomy defect manifesting as irregularity of the right posterior 6th rib on the frontal radiograph) at least 8 years earlier for a "right lung cyst," which was reportedly benign but incompletely resected due to close contact with the pericardium posteriorly, but no further information was available.

Diagnosis: Bronchogenic cyst, recurrent, due to incomplete previous resection

References

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